Diagnostic and Interventional Imaging (2015) **96**, 297—299

LETTER / *Thoracic imaging*

About chemodectomas occurring concurrently in two sites: In the mediastinum and the lungs

Keywords: Chemodectoma; Imaging; Dual location; Mediastinal; Pulmonary

Case report

The patient is a 48-year-old female patient with an unremarkable medical history who underwent a lung X-ray for bronchitis. The lung X-ray showed overhanging of the mediastinal left aortic arch suggestive of a mediastinal mass. A chest CT scan was carried out with a slice thickness of 0.75 mm and mediastinal and parenchymal window settings, with and without contrast injection for enhancement of the arterial and venous phases. The CT scan showed two well-delimited masses with regular contours, one located in the anterosuperior mediastinum measuring 35 × 27 mm (Fig. 1) and the other in the apical segment of the parenchyma measuring 15 mm (Fig. 2). The two tumours showed intense enhancement early after contrast injection, otherwise no mediastinal adenomegaly or other thoracic lesions were observed.

The conclusion after the scan was that the tumour was a hypervascular tumour with a double location (mediastinum and lung) and the first hypotheses pointed to the diagnosis: a paraganglioma, a carcinoid tumour or a thymoma...

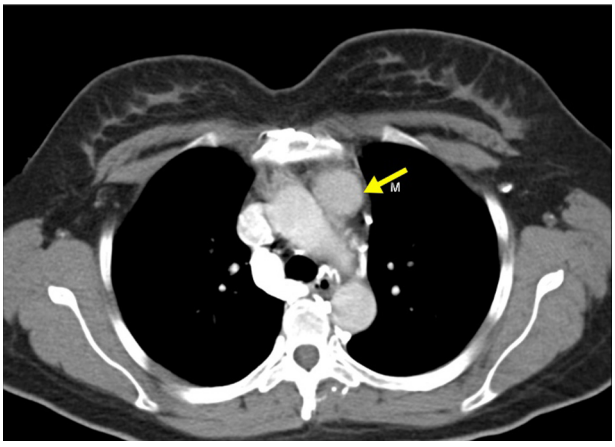

Figure 1. Small, well-delimited, homogeneous anterosuperior mediastinal mass enhanced following contrast injection, in front of the aortic arch and behind the sternum with no lytic bony lesions.

Histopathological examination of samples obtained by CT-guided biopsy of the anterior mediastinal mass confirmed the diagnosis of a benign, non-secretory chemodectoma-type paraganglioma.

Monitoring was indicated for the patient in the absence of a compressive syndrome or symptoms of heart disease or hypertension.

Discussion

Paragangliomas are tumours that develop from the adrenal medulla (pheochromocytomas) or in the extra-adrenal paraganglia. Only extra-adrenal paraganglia will be discussed in this article. Paraganglionic tissue (neuroendocrine system) is made up of cells deriving from the neural crest and may be divided into two groups.

Paraganglionic tissue from the *first group* is distributed along the length of the pre- and para-vertebral sympathetic chain and the sympathetic nervous system innervating the pelvic and retroperitoneal organs (orthosympathetic system: retroperitoneal chromaffin cells including the organ of Zuckerkandl) and gives rise to retroperitoneal paragangliomas, mediastinal paragangliomas, spinal paragangliomas, filum terminale, cauda equine, paragangliomas of the kidneys, bladder, endometrium, vulva, heart and lungs [1—3].

Paraganglionic tissue from the *second group*, distributed along the length of the parasympathetic nervous system and concerning mostly the cervical and thoracic branches of the glossopharyngeal and pneumogastric nerves (parasympathetic system: nonchromaffin cells with a chemoreceptor function: head, neck, mediastinum), gives rise to paragangliomas of the head and neck (chemodectomas): carotid paragangliomas, jugular-tympanic paragangliomas, vagal paragangliomas, paragangliomas of the orbits, the nasal fossa, the nasopharynx, the trachea, the thyroid and the larynx. They are never located on the limbs [1—3].

Paragangliomas are mostly sporadic, single tumours, occurring in subjects between 40 and 60 years of age. However, the tumours may develop at multiple sites and familial forms also exist but in such cases, the patients are younger. They are sometimes combined with a multiple endocrine neoplasia syndrome. Paragangliomas are discontinuous aggregates of neural crest cells. They are found in the adrenal medulla, along the aorta, in blood vessel walls

http://dx.doi.org/10.1016/j.diii.2014.11.006

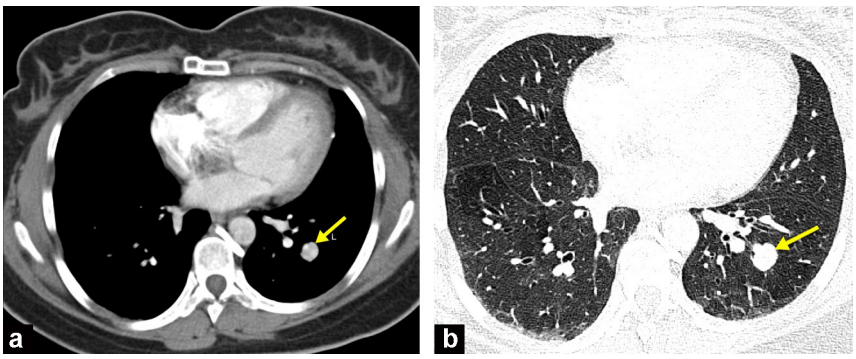

Figure 2. Small lung tissue lesion with the same characteristics as segment 6 of the inferior lobe of the left lung; axial slice, mediastinal window (a) and parenchymal window (b) showing its perivascular location.

and in certain organs such as the heart, the prostate or the ovaries. Paragangliomas develop at the cost of paraganglionic structures. Tumours that develop from sympathetic structures are most often active and are referred to as pheochromocytomas in the adrenal glands and active paragangliomas when they develop outside the adrenal glands. Tumours that develop from parasympathetic structures are mostly inactive and referred to as chemodectomas or inactive paragangliomas. Thoracic paragangliomas account for less than 2% of mainly adrenal paragangliomas [4] and their concurrent occurrence in two sites is exceptional.

Macroscopically, chemodectomas are soft and richly vascularised tumours. Histologically, the cells are oval, separated by reticulin fibres and show no mitotic activity. However, distinguishing chemodectomas from pheochromocytomas may be tricky [4]. Currently, the distinction between secretory and non-secretory tumours is based on laboratory findings and not on histopathological examination alone. Only 10% of pheochromocytomas, but 20% of the extra-adrenal forms, are malignant. Malignancy is based on the presence of metastases.

Paragangliomas occur most frequently in adults between 30 and 40 years of age. Chemodectomas may also present as a mass leading to the compression of an intercostal nerve, the spinal cord, Claude Bernard-Horner syndrome [5] and even symptoms of heart disease due to compression of the coronary artery or atrium [6]. The symptoms of pheochromocytomas are mainly related to their secretion of catecholamines giving rise to hypertension, stroke, renal or cardiac insufficiency, weight loss, diabetes, etc. More atypical symptoms such as tachycardia, anxiety, palpitations, nausea, vomiting or headache may also be observed. Paragangliomas may sometimes secrete other hormones, giving rise to Cushing's syndrome (secretion of ACTH), polycythemia (secretion of EPO), hyperkalcemia (secretion of PTH) and diarrhoea (secretion of VIP).

Malignant paragangliomas may be immediately metastatic with a frequency estimated at 20%. Bone metastases are the most common metastases. Other sites of metastasis include regional lymph nodes, the lung and the liver. Skin and brain metastases have been reported more rarely [4]. In close to 10% of cases, pheochromocytomas are associated with multiple endocrine neoplasias (MEN IIa and IIb), Von Hippel-Lindau disease or neurofibromatosis [4]. In some cases, several paraganglionic tumours may be observed concurrently which should call to mind the Carney triad (coexistence of multiple extra-adrenal pheochromocytomas, lung hamartomas and gastric gastrointestinal stromal tumours) [7].

Imaging techniques are used to precisely locate the tumour and determine the extent to which it has spread. The concurrent occurrence of chemodectomas in two sites has rarely been described. Paragangliomas may be observed in the cardiac field and in the pulmonary grooves [8]. A chest X-ray may show enlargement of the superior mediastinum, a change in the cardiac silhouette or lung metastases.

A CT scan without and then with contrast injection can be used to better visualise the lesion and specifically analyse the tumour by defining its morphological characteristics and its topography. The tumour is generally a homogeneous, well-delimited tissue mass isodense to muscles showing intense enhancement after contrast injection. CT scans also enable the study of the tumour with respect to different organs as well as tumour extension. However, due to the proximity of the heart and large blood vessels, cardiac gating may be required for small tumours not to be missed. With MRI, the tumour is isointense to muscles, hyperintense on T2-weighted images with a serpiginous appearance indicative of vascular involvement. The salt and pepper appearance due to the juxtaposition of hypersignals and hyposignals is not always seen, but contrast uptake is consistently massive.

Diagnostic arteriography is an optional test used to visualise the tumour blush characteristic of hypervascular tumours and to map blood flow. A further advantage of the test is that it can be used for pre-surgical embolization to reduce preoperative bleeding and the volume of the tumour.

The sensitivity and specificity of MIBG scintigraphy is respectively 70% and 98% for the diagnosis of paragangliomas [9], irrespective of whether they are secretory or not. Possible alternatives include somatostatin analogue scintigraphy (OctreoScan) that displays a sensitivity of 22% [10], or PET with 6-(18)F-fluorodopamine, found to be the most sensitive technique (90%) in a recent study [10]. Nonetheless, OctreoScans are still useful in cases of metastatic or extra-adrenal paragangliomas.

Tumour appearance with medical imaging is not specific, particularly when the tumours occur in unusual locations like in this case. Consequently, it is important to make a diagnosis so anaesthetic accidents may be avoided during

surgery and to implement special procedures for general anaesthesia.

Localised paragangliomas are treated surgically. Prior to surgery, patients may require embolization because non-secretory tumours are often richly vascularised. Surgery is performed using a direct approach by sternotomy or thoracotomy depending on where the tumour is located. Extracorporeal circulation may be required for the removal of infiltrating tumours [5]. Complete tumour resection is curative in cases of localised tumours [5]. In cases of malignant disseminated tumours, chemotherapy or Iodine-131 MIBG scintigraphy only helps to stabilise the disease and progression generally occurs within 2 years.

Conclusion

Chemodectomas are tumours of the extra-adrenal paraganglionic system located along the length of the cervical-cranial parasympathetic nervous system. Tumour occurrence in the mediastinal area is rare and a double mediastinal and lung location is even rarer. CT scans and MRI are essential for making a diagnosis and studying the tumour with respect to the other organs but the information acquired by these imaging techniques is unspecific, particularly when the tumours occur in unusual locations such as in this case. Consequently, it is important to make a diagnosis so anaesthetic accidents may be avoided during surgery and to implement special procedures for general anaesthesia.

Disclosure of interest

The authors have not supplied their declaration of conflict of interest.

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